Keratoconus is an ocular disorder characterized by progressive corneal thinning and other corneal sequelae, in which the cornea assumes an irregular conical shape. It can be classified according to degree of conicity and characterized morphologically by the shape of the cone. Although early keratoconus with minimal irregular corneal astigmatism may be corrected by spectacles, they cannot be expected to improve visual acuity to the same extent that rigid contact lenses offer, when irregular astigmatism is present or increases.

**DIAGNOSIS**

Since the onset of this disorder usually occurs in the second decade of life, these patients are active occupationally, socially and recreationally. Therefore demands for good visual acuity among these patients, is a major concern for both the practitioner and the patient. Keratoconus quite often typically progresses for a period of five to seven years at a variable rate. The end point of this progression may range from slight corneal irregularity to severe distortion of the corneal contour and apical scarring. Some of these cases require surgical keratoplasty intervention if the patient cannot achieve acceptable vision with spectacles or is unable to tolerate contact lenses. The most practitioners can do for these patients is offer the best visual acuity possible until this condition reaches some point of remission or until surgical intervention is required.

Since the introduction of photokeratoscopy and, most recently, corneal molding, much has been learned regarding the corneal topographical changes the cornea undergoes during the early stages of keratoconus.

1. The inferior portion of the cornea steepens.
2. The corneal apex steepens and assumes an oblique to “against-the rule” astigmatic configuration.
3. The superior cornea (above the horizontal midline) remains relatively normal and unaffected.

As this condition progresses, the curvature of the corneal apex becomes more elongated and the corneal surface bulges more creating increased irregular astigmatism. For these reasons, acceptable visual acuity with eyeglasses becomes increasingly impossible. Soft lenses, due to their flexible nature and limited ability to correct corneal astigmatism, do not provide the degree of visual acuity these young, active patients require to function in their daily lives. By their physical nature, rigid contact lenses offer the best vehicle for providing very good visual acuity to these patients, until their condition reaches a point of remission or until surgical intervention is indicated. These patients typically become more dependent...
on their rigid contact lenses for day and night time vision correction. It is therefore imperative to maximize lens-wearing comfort for the patient. Because of the prolonged wearing periods, it is also critical to protect the already compromised integrity of the corneal physiology. The use of gas permeable lens materials offering high oxygen transmissibility is extremely important.

Because of the limited and often inaccurate information provided by keratometry, keratoconus lens fitting dictates that diagnostic lenses are used to achieve the best possible fit.

**CLASSIFICATION**

Mild cones may be classified as having keratometry ("K") readings in both meridians of <48.00 diopters, moderate cones having "K" readings of 48.00 to 54.00 diopters and severe cones as having "K" readings of >54.00 diopters. Since the superior portion of the cornea is relatively normal, it is an important consideration when selecting a lens base curve to avoid impingement in this area.

**IDENTIFICATION OF CONE TYPE**

It is also helpful for the fitter to be able to identify the shape and size of the cone they are dealing with, to be able to help assist with diagnostic lens selection, minimize the amount of lens parameter changes and subsequent chair time involved in fitting these cases.

The contact lens fitter can determine the type and size of the cone in one of several ways. Following dilation of the pupils, the "red reflex" can be used to retro illuminate the cornea and determine the shape and approximate size of the cone. A more direct approach is to place a flat-fitting lens on the eye and view the outline produced by the flat fitting relationship as viewed using sodium fluorescein.

1. <5mm for “nipple” cones—usually located at or near the center of the corneal apex.
2. 5mm to 6mm for “oval” cones. These cones are typically large in size and ellipsoidal in shape. They are located predominantly inferior-temporally and appear to sag due to their larger horizontal dimension.
3. >6mm for “globus” cones—these cones appear more diffuse and are of larger size. They may extend above the horizontal midline of the cornea. As progression of the disease is noted and corneal thinning continues, Fleischer’s ring hydrops, positive Munson’s Sigh striae and central scarring are among the signs that may be seen.

**RIGID LENS FITTING**

While keratoconus can present numerous lens-fitting difficulties, these can be minimized if a disciplined approach to lens fitting is used. In the early stages of this condition, use of rigid spherical and aspheric lenses employing steep base curves, standard designs and average lens diameters will function very well to provide good patient comfort and very good visual acuity.

As the disease progresses, fitting moderate to severe keratoconus requires more attention to identifying the size and shape of the corneal aberration. As stated previously, use of diagnostic lenses is the only sure method to achieve an optimal lens fitting relationship. Basic lens design requirements dictate that lenses provide steep base curves, relatively small posterior optical zones and flat peripheral curves. There are several design systems that meet these criteria.

**MCGUIRE CONE LENS DESIGN**

The McGuire fitting philosophy indicates a “three-point touch” fitting system. The goal is to achieve superior alignment between the cornea and the lens. The ideal fit appearance will exhibit 2mm to 4mm of “feather” apical bearing, with slight edge lift inferiority.

Attempts to vault the corneal apex to reduce inferior edge lift will result in an excessively steep fitting relationship on the more normal superior portion of the cornea.

The fitting system consists of three separate diagnostic lens sets specifically designed for each type of cone configuration. The design is predicated on the lens optical zone size as it relates to the size of the cone varying form 6mm for the nipple-type cones to 6.5mm for oval cones and 7mm for globus cones. Each incorporates identical peripheral curve systems blended together to resemble an aspheric-like flattening in the posterior lens periphery. Four peripheral curves are utilized:

- 0.5mm flatter than the base curve
- 1.5mm flatter than the base curve
- 3.0mm flatter than the base curve
- 5.0mm flatter than the base curve

**Soper Keratoconus Design**

The Soper keratoconus design utilizes 10 base curves and standard optical zones sizes identified by degree of cone severity. Letters identify these lenses. They indicate the type of cone for which they might be suited. The system recommends vaulting
Use of Contact Lenses in the Visual Correction of Keratoconus

When talking to patients, consider the following, “republished with permission of AllAboutVision.com.” This material was updated Dec. 2008.

KERATOCONUS
By Gretchen Bailey and Judith Lee; additional contributions and review by Gary Heiting, OD

Often appearing in the teens or early twenties, keratoconus is a progressive eye disease in which the normally round cornea thins and begins to bulge into a cone-like shape. This cone shape deflects light as it enters the eye on its way to the light-sensitive retina, causing distorted vision. Keratoconus can occur in one or both eyes.

Keratoconus Symptoms and Signs
Keratoconus can be difficult to detect, because it usually develops so slowly. However, in some cases, it may proceed rapidly. As the cornea becomes more irregular in shape, it causes progressive nearsightedness and irregular astigmatism to develop, creating additional problems with distorted and blurred vision. Glare and light sensitivity also may be noticed. Keratoconic patients often have prescription changes every time they visit their eyecare practitioner. It’s not unusual to have a delayed diagnosis of keratoconus, if the practitioner is not familiar with the early-stage symptoms of the disease.

What Causes Keratoconus?
New research suggests the weakening of the corneal tissue that leads to keratoconus may be due to an imbalance of enzymes within the cornea. This imbalance makes the cornea more susceptible to oxidative damage from compounds called free radicals, causing it to weaken and bulge forward.

Risk factors for oxidative damage and weakening of the cornea include a genetic predisposition, explaining why keratoconus often affects more than one member of the same family. Keratoconus is also associated with overexposure to ultraviolet rays from the sun, excessive eye rubbing, a history of poorly fit contact lenses and chronic eye irritation.

Keratoconus Treatment
In the mildest form of keratoconus, eyeglasses or soft contact lenses may help. But as the disease progresses and the cornea thins and becomes increasingly more irregular in shape, glasses or soft contacts no longer provide adequate vision correction.

Treatments for moderate and advanced keratoconus include:
• Gas permeable contact lenses: If eyeglasses or soft contact lenses cannot control keratoconus, then rigid gas permeable (RGP) contact lenses are usually the preferred treatment. Their rigid lens material enables RGP lenses to vault over the cornea, replacing the cornea’s irregular shape with a smooth, uniform refracting surface to improve vision. But RGP contact lenses can be less comfortable to wear than a soft lens. Also, fitting contact lenses on a keratoconic cornea is challenging and time-consuming. You can expect frequent return visits to fine-tune the fit and prescription, especially if the keratoconus continues to progress.

• Hybrid contact lenses: Hybrid contact lenses have a relatively new design that combines a highly oxygen-permeable rigid center with a soft peripheral “skirt.” Manufacturers of these lenses claim hybrid contacts provide the crisp optics of a GP lens and wearing comfort that rivals that of soft contact lenses. Hybrid lenses are also available in a wide variety of parameters to provide a fit that conforms well to the irregular shape of a keratoconic eye.

• Intacs: (Addition Technology, Des Plaines, Ill.) Intacs or corneal inserts received FDA approval for treating keratoconus in August 2004. These tiny plastic inserts are placed just under the eye’s surface in the periphery of the cornea and help reshape the cornea for clearer vision. Intacs may be needed when keratoconus patients no longer can obtain functional vision with contact lenses or eyeglasses.

• Boston Scleral Lens Prosthetic Device (BSLPD): This cone-shaped device resembles a large contact lens and works partly by maintaining a “pool” of fluid on the eye’s surface through which light rays pass and are bent to achieve proper focus. Rather than resting on the eye’s clear surface or cornea, however, the lens makes contact with the “white” or sclera of the eye. The device was FDA-approved in 1994.

• C3-R: (Boxer Wachler Vision, Los Angeles) Another new procedure for treating keratoconus, known by the brand name of C3-R (corneal collagen cross-linking with riboflavin), is a non-invasive method of strengthening corneal tissue to halt bulging of the eye’s surface. In the C3-R procedure, eye drops containing riboflavin (vitamin B2) are placed on the cornea and are then activated by ultraviolet (UV) light to strengthen links between the connective tissue (collagen) fibers within the cornea.

• Corneal transplant: Some people with keratoconus can’t tolerate a rigid contact lens, or they reach the point where contact lenses or other therapies no longer provide acceptable vision. The last remedy to be considered may be a cornea transplant, also called a penetrating keratoplasty (PK or PKP). Even after a transplant, you most likely will need glasses or contact lenses for clear vision.
the corneal apex. The theory behind this fitting system (Korb, et al) postulates that apical bearing should not be used because of the possibility of corneal complications including abrasions and accelerated corneal scarring.

1) Mild — (A, B, C, D) less than 48 diopters in either corneal meridian.
2) Moderate — (E, F, G) 48 to 54 diopters in either corneal meridian.
3) Advanced — (H, I, J) >54 diopters in either corneal meridian.

The difference in this fitting system is that overall lens diameter and base curve radius is changed to increase or decrease lens sagittal depth, thereby allowing this design to be used in fitting a range of corneal steepness. Letters to signify which type of cone each would fit identifies lenses.

ASPHERIC RIGID LENS DESIGNS
In keratoconus, aspheric designs have proven to not only fit the varied corneal shapes, but also simplify the fitting process by eliminating some of the variables involved with lens design and fitting. These designs provide improved mid-peripheral alignment as compared to spherical designs. Optical zone sizes are generally small but will usually accommodate the larger globus cones as well as oval or nipple-type ones.

Several of these aspheric designs are also “junctionless.” That is, the design has been mathematically calculated to allow the various aspheric curves to be joined in a matter in which no junctions are created. This helps to eliminate narrow bearing areas in the mid-periphery and allows lens mass to be distributed more evenly, creating a more uniform tear layer profile under the lens.

OTHER CONTACT LENS OPTIONS
Other means of fitting the keratoconic patient have also been utilized. A “piggyback” fitting systems has been used in cases where corneal contact lenses will not suitably stabilize on the cornea, where rigid lens wear alone is not tolerated by the patient or metabolic problems exist. Using this technique, the patient is first fitted with a large, thin soft lens in the conventional manner so that both good lens centration and movement are achieved. Once the patient has adapted to full-time soft lens wear, keratometry readings are taken of this “renewed” corneal surface and rigid lens diagnostic fitting is carried out. Use of high-permeability rigid lens materials, designed as thin as possible is essential, since the patient is wearing a “double layer” of lenses on the cornea and the physiology is already impaired. The disadvantage of this system is that the patient must contend with insertion, handling and caring for both types of lenses separately. Use of separate care systems is also costly and time-consuming.

Another option is the use of a hybrid lens such as the SoftPerm lens. The advantage of this system is that the soft lens and gas permeable lenses are polymerized together into one unit.

Consideration must be given to the physiological impact that these fitting systems may have on the cornea with regard to providing adequate oxygen.

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